

Burnett (S. M.)
History of case and p. m. examinations
the latter made Sep. 11. 1895. by Dr. D. L.
Harris Case of Dr. S. M. Burnett.
Specimens of brain + inner meninges
to Dr. Burnett.

Double exophthalmos x x x x

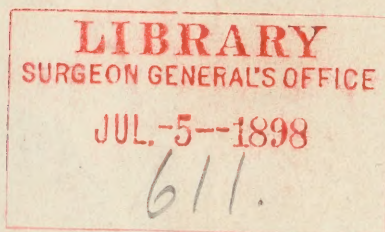




FIG. 1



FIG. 2

DOUBLE EXOPHTHALMOS WITH ULCERATIVE DESTRUCTION OF THE EYES IN AN INFANT FROM SARCOMA OF THE DURA MATER, STARTING MOST PROBABLY AT THE SELLA TURCICA—EXTENSIVE PATHOLOGIC CHANGES IN THE BONES—DEATH—AUTOPSY.

By SWAN M. BURNETT, M.D., PH.D.

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Jessie Irving, a well-nourished mulatto infant of five months, was brought to my clinic, at the Emergency Hospital, by her mother, with the statement that she had first noticed something queer about her right eye some two months before. At the time of examination, August, 1893, there was a marked exophthalmos of the right eye, which was turned strongly outward. There was paralysis of all the muscles of that eye except the external rectus. The child was too young to determine positively whether there was any sight remaining. The fundus appeared unchanged from the normal in any important particular. In addition to the eye trouble there was a diffuse swelling in the temporal region of the right side. The girl was born in a normal labor, without the assistance of any instruments, and there was no history in either father or mother of a syphilitic taint. Up to that time the child had been perfectly healthy, and even now made no complaint, and took the breast and slept well. The swelling in the temporal region gradually enlarged, and at the end of a month became soft at a point midway between the upper border of the auditory canal and the external angle of the eye. An exploratory incision was made under chloroform, and a quantity—estimated at two ounces—of brownish yellow liquid escaped. A probe was introduced and disclosed a cavity inside the cranium which was probably the size of a large walnut. Following this probing there was an excessive hemorrhage which could only be controlled by packing the cavity with

iodoform gauze and applying a tight bandage. The child was taken into the hospital and at the end of four days the dressings were removed. There was no further hemorrhage. The cavity was packed again with iodoform gauze, and this was repeated at proper intervals until the cavity closed at the end of some three months. The child, in the meantime, continued in perfect health, and was bright and cheerful. The exophthalmos quite distinctly diminished, but the motion of the eyeball continued as restricted as before. There was at that time no appearance of change in the nerve head, but it was doubtful whether any vision remained in the eye.

The patient then disappeared from observation and was not seen again until the spring of 1895. It was then as well grown as any child of that age, and was in perfect health. The right eye was still somewhat more prominent than the left, and the paralysis had persisted. Examination showed that the wound in the temporal region had healed perfectly, though that side was somewhat more prominent than the other. The great amelioration in the condition was attributed to prayers.

Some weeks afterwards the child was again brought to the clinic with an increase in the exophthalmos which had been noted as beginning soon after the last visit. It was, however, unattended with any general disturbance. This exophthalmos increased from week to week. Suddenly one morning in June, 1895, it was discovered by the parents that the child could not see out of the other (left) eye, though it had been playing around the room as usual the night before. The child was now totally blind. An examination revealed no change in that eye either externally or internally. There were not then, nor had there been at any time, symptoms of paralysis of any of the cerebral or spinal nerves, other than those mentioned. Soon an exophthalmos began to be noticed in the left eye also, and it was thought best to send her to the Children's Hospital.

The following notes of the case during her stay in the hospital are kindly furnished by the Resident Physician, Dr. Ashford:

"J. I., aged two years and four months, admitted July 12, 1895. Family history good; just recovering from measles; well

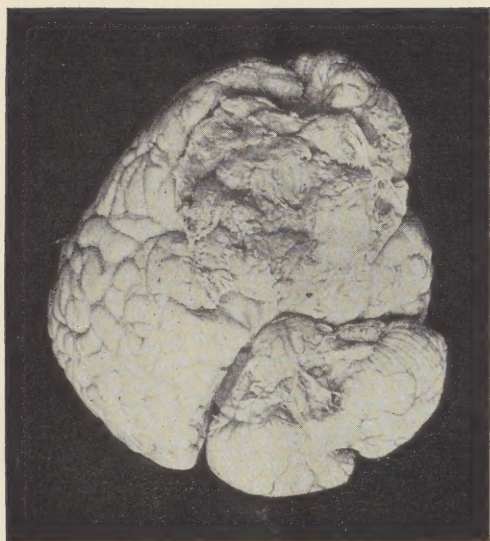


FIG. 3



FIG. 4

nourished. Both eyes exophthalmic, right much more so than left. Right eye immobile and there is sloughing of the cornea. Left eye nearly fixed, but some movement of the lids. Child listless, but no paralysis anywhere except in the eyes. Appetite good; bowels good; sleeps well. There is a discharge of thick purulent matter, mingled with blood, from the right eye and the right nostril, and some purulent discharge from the left nostril. August 5th — exophthalmos of both sides much increased, and there is a prominence at the root of the nose on the right side in which there is a sinus connecting with the post-nasal cavity, and through this there is a discharge of very offensive matter. The upper lid of the left eye is still movable, but both eyeballs are fixed. Ulceration of right cornea increasing. The appetite is still good, and she sleeps well; there is, however, a distinct loss of flesh; no pain complained of (Photograph, Fig. 1, was taken at this time). August 12th — increase in the swelling at root of nose, and the left cornea is beginning to ulcerate. Though she eats and sleeps well, she is getting more emaciated and weaker. The condition steadily grew worse from this time on. On Sept. 5th, the pulse was weak, slight twitching of the muscles of the right hand during the night; left side colder than right. On September 11th, she died quietly at 5.30 A. M." (Photograph, Fig. 2, was taken by Dr. Gray after death).

An autopsy was made by Dr. D. S. Lamb of the Army Medical Museum, that same afternoon. The following is an account of the

POST MORTEM EXAMINATION.

"Emaciated. Purpuric spots over body. Consistency of skull good. Sutures well united. Dura mater very adherent. Abundant cerebral fluid. Partial absorption of frontal lobe of brain on right side. Mass attached to left orbital plate and can be scraped off from before backwards. Growth began outside the dura mater, and extended backwards. Dura thickened back of tumor. Brain not involved in any place. Pus on right side in olfactory region. Ethmoid seems to be entirely destroyed. Large mass of new bone affecting right orbital plate. It is twelve times thicker than normal. Left orbital plate pushed upward and four times thicker than normal from new bone.

Part of contents of left orbit taken out for examination. Whole posterior portion of orbital side of sphenoidal fissure destroyed on right side. Wings of sphenoid in good condition except at base. Nasal bones are disarticulated and pushed to left side. Nasal fossæ filled with hypertrophied tissue and greenish pus. At junction of parietal, malar, sphenoid, and frontal bones there is excessive hypertrophy. Greater wing of sphenoid hypertrophied so as to nearly obliterate the middle fossa of the skull. Brain apparently normal. Tumor is flat, two and three-quarter inches long, two and one-half inches broad, one and one-half inches thick at thickest part (Fig. 3 shows the relative size and position of the tumor at the base of the brain). It is vascular, reddish, limited on brain side by dura, and has points of degeneration."

It could be easily seen on inspection of the specimen after its removal from the cranial cavity that the optic nerves pierced the dura mater and passed out into the substance of the tumor at a distance of one and one-half centimeters from the commissure on the right side and two centimeters on the left side. The third nerve passed through the dura and into the tumor some distance behind this. From these appearances, in connection with the clinical history, we can infer very justly, I think, that the morbid process began under the dura on the right side near the anterior clinoid process, since it involved the third nerve first of all, but anterior to the posterior clinoid process, since the sixth nerve was not affected. The optic nerve was probably not implicated at that time to any serious extent. What was the nature of the pathologic change at the beginning, which, while evidently so limited as to the central parts, was sufficient to cause such extensive manifestations in the right temporal region and orbit? It would seem most likely that it was of a cystic character, or, at least, that there was an unusual amount of fluid formed, the pressure of which was exerted in this particular direction. When this was evacuated and drainage was established, the morbid process was checked, or held in abeyance for at least eighteen months, at any rate so far as the fluid formation was concerned. The original lesion, however, still persisted and was extending forward

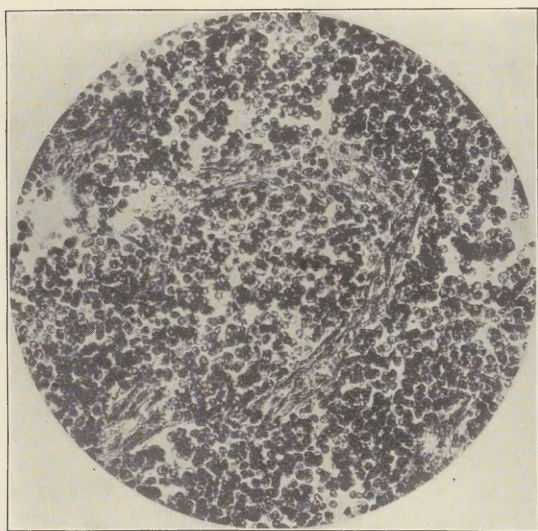


FIG. 5

in a manner to involve more slowly the bones on the right side of the sella turcica. That it did not extend over to the left side until much later is evidenced by the sudden appearance of blindness of the left eye, indicating an involvement of that nerve, and before the third nerve on that side was affected, only a short while before the end. The very early appearance of the trouble, about the second or third month after birth, would lead us to suspect that the morbid process had its origin *in utero*, and was congenital. Another interesting feature, from a general pathologic point of view, is that the primary changes were limited, almost altogether, to the bony tissue, these changes being both of a destructive and hypertrophic character (Fig. 4 shows a horizontal section through both orbits and sphenoids). Though lying under and most probably springing from the dura mater, it nowhere perforated that membrane. The histology of the mass showed it to be a sarcoma (Fig. 5). The disease had not, however, been carried by metastasis to any organs or tissue at a distance, but remained strictly localized, although the seeds of the disease had been present in the system ever since birth, and probably before.

DISCUSSION.

DR. J. O. TANSLEY, New York. I have had several of these cases of pulsating exophthalmos, two of which stand out in my memory and act as beacons in telling me what to do in similar cases. One of them I reported to this Society a few years ago, and I will give a short synopsis of the second.

A doctor of medicine came to New York to study ophthalmology and otology and fell into my hands. He had a very slight discharge from his nose, and a small piece of ill-smelling sputa would be expectorated from the naso-pharynx each morning, but at no other time. He was advised to pay no attention to it. He was going to London and took letters of introduction to Sir Morrill McKenzie, whom he consulted about his condition. Dr. McKenzie told him that he had ethmoid disease and he would not operate upon him, but recommended him to one of his friends. He accepted this advice and I have a written description of the operation. The doctor made an examination with a probe, said that he distinctly felt dead bone which he seized with a pair of forceps and exerted some traction, which was almost immediately followed by a gush of blood that filled

the whole nasal cavity, and, in a few moments, the patient, the bed, and the doctors were deluged in blood. They stopped the operation, of course, filled the nose with strips of gauze that were packed very strongly in the nasal cavity, and happily stopped the hemorrhage. But almost immediately the left eye began to bulge, and continued to do so, until it protruded markedly from the orbit; pulsating also. When the patient came out of ether, he complained of a disagreeable bruit in his head. This was listened for, and, by placing the ear on the head, could be distinctly heard. They put him on a very low diet, with a limited supply of liquids, so that he was almost starved to death. They ligated the left carotid and the pulsation and bruit disappeared, but the exophthalmos continued, and the pulsation was resumed in four days after the ligation. Of course, they expected him to die, and they made every arrangement to ship his body to this country. After a number of weeks the patient became restless, and said he would rather "Eat, drink, and be merry," and die, than to suffer from hunger and thirst, and live miserably. The exophthalmos gradually disappeared, but the bruit continued for a number of months. This same physician was an assistant in my clinic for a year, and is now practicing ophthalmology and otology in one of the cities of Canada. He has, of course, no sight in the left eye. He is very happy in having an excellent practice, and it may be a pleasure to you, as it is to me, to know that he can tell as good and as many stories as our friend John Green.

